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FIT Clinical Decision Making

RESTRICTIVE CARDIOMYOPATHY DUE TO ATRIAL NATRIURETIC FACTOR TYPE AMYLOIDOSIS

Poster Contributions
Hall C
Sunday, March 30, 2014, 9:45 a.m.-10:30 a.m.

Session Title: FIT Clinical Decision Making: Heart Failure / Cardiomyopathies
Abstract Category: Heart Failure and Cardiomyopathies
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Background: Atrial natriuretic factor type of amyloid has been responsible for development of isolated atrial amyloidosis. Although clinical significance of this type of amyloid is controversial, it may play a role in development of significant cardiac functional abnormalities.

Case: A 72-year old Caucasian female presented with three months history of progressive shortness of breath. This started after she underwent atrioventricular node ablation and pacemaker placement for permanent atrial fibrillation. She was readmitted, transthoracic echocardiogram (TTE) showed moderate pericardial effusion and she underwent pericardiocentesis. Later she developed bilateral pleural effusion that worsened progressively to the point of having recurrent thoracocentesis. The shortness of breath continued to worsen and she also developed bilateral lower leg edema and ascites.

Decision Making: She was then admitted to St. Luke's hospital. On examination, she had raised Jugular venous pressure, positive Kussmaul sign, grade 3/6 systolic murmur in the mitral and tricuspid areas, distended abdomen, and bilateral leg edema. TTE showed mild systolic dysfunction, grade 3-4 diastolic dysfunction, moderate mitral regurgitation, and severe tricuspid regurgitation. Right and left heart catheterization showed equalization of right and left sided end diastolic pressure at 15mmHg without evidence of ventricular discordance, consistent with restrictive physiology. Catheterization also showed triple vessel disease. She underwent coronary artery bypass grafting, mitral, tricuspid valve annuloplasty, and left atrial appendage (LAA) excision. Congo red stain performed on LAA specimen was consistent with amyloidosis. The block was sent to Mayo clinic for amyloid subtyping. According to their report, LAA specimen was involved by amyloidosis, alpha atrial natriuretic factor (AANF) type. Therefore, diagnosis of restrictive cardiomyopathy due to AANF type of amyloidosis was established.

Conclusion: Since incidence of AANF type of amyloid is quite high in surgically resected left atrium, its clinical significance is neglected. It should be considered in patients with unexplained heart failure.